

Health Care Provider Fact Sheet

Disease Name

Sickle Cell Disease

Alternate Name(s)

Sickle Cell Anemia

Acronym

Hb S/S

Disease Classification

Hemoglobinopathy

Symptom onset

May be asymptomatic.

Symptoms

Any sign of illness in an infant with sickling disease is a potential medical emergency. Acute and chronic tissue injury can occur when sickled cells cause vascular occlusion. Sickling diseases can cause severe pain anywhere in the body, but most often in the hands, arms, chest, legs and feet. Complications may include, but are not limited to, the following: sepsis, acute chest syndrome, hand-and-foot syndrome, splenic sequestration crisis, aplastic crisis, stroke and painful episodes.

Natural history without treatment

Infants with sickle cell disease are vulnerable to serious bacterial infections that can be life threatening.

Natural history with treatment

Reduced mortality and morbidity with penicillin prophylaxis.

Treatment

The National Institutes of Health clinical guidelines for management of sickle cell disease state, "Penicillin prophylaxis should begin by 2 months of age for infants with suspected sickle cell anemia, whether or not the definitive diagnosis has been established." Antibiotic therapy should continue until at least 5 years of age.

Emergency Medical Treatment

See sheet from American College of Medical Genetics (attached) or for more information, go to website:
http://www.acmg.net/StaticContent/ACT/ACT_sheet_HBSS_FS__4.28.06%20ljo.pdf

Inheritance

Autosomal recessive

General population incidence

1:500 for African American and Hispanic American.

OMIM Link

<http://www.ncbi.nlm.nih.gov/entrez/dispomim.cgi?id=603903>

Genetests Link

www.geneclinics.org

Support Group

Sickle Cell Information Center
<http://www.scinfo.org/>

Sickle Cell Disease Association of America, Inc.
<http://www.sicklecelldisease.org>

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